was significantly higher than in groups 1,2 ($p < 0,01$ and $p < 0,04$ accordingly) at the end of the study.

Conclusion: Mean values of EScSG-AI score associated with and were accompanied by progression pulmonary alterations in pts SSc so EScSG-AI score can be used as valuable tool for long-term follow-up studies.

Disclosure of Interests: None declared

DOI: 10.1136/annrheumdis-2019-eular.6818

AB0218 CORRELATION BETWEEN IMMUNOLOGICAL PARAMETERS AND PULMONARY FUNCTION PARAMETERS IN THE PATIENTS WITH SSC AND ILD OVER 5 YEARFOLLOW UP STUDY

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Background: Interstitial lung disease (ILD) is related to specific radiographic features in lung imaging and/or the presence of restrictive disorders in pulmonary function tests (PFTs). ILD is one of the leading causes of death in systemic sclerosis patients. Major risk factors of ILD associated with SSc (SSc-ILD) include male sex, diffuse type of cutaneous SSc and presence of anti-Scl-70 antibodies.

Objectives: Taking into account that anti-Scl-70 antibody is an unfavorable predictor of ILD, we have assessed the time course of FVC and DLco in anti-Scl-70-positive and anti-Scl-70-negative patients.

Methods: It was a longitudinal study involving 77 pts with SSc-ILD (mean age was $46,2 \pm 13,4$; 69% have limited subset of the disease; 93% were female). The mean duration of follow up was $58,9 \pm 11,4$ months. Pts. were investigated with HRCT twice (at first visit and at the end of the study) and according the CT-changes were divided into 3 groups: the 1st group (16 pts) with improvement; 2nd group (39 pts) without any changes and 3rd group (22 pts) with worsening of fibrosis. We evaluated the forced vital capacity (FVC), diffusing capacity of carbon monoxide (DLCO) in one year and in 5 years and anti-Scl-70 antibodies at the end of the study.

Results: In anti-Scl-70-negative patients the average FVC remained unchanged within a year ($91,4 \pm 17,4\%$ и $91,6 \pm 17\%$, respectively), however, within 5 years FVC significantly increased to $99,1 \pm 20\%$ ($p < 0,001$), while in anti-Scl-70-positive patients the average FVC was lower at the baseline and remained virtually unchanged both 1 year and 5 years later ($85,4 \pm 17,3\%$. $85,5 \pm 17$ и $86 \pm 21\%$, respectively), as a result in 5 years' time the average FVC in anti-Scl-70-negative patients was significantly higher than in anti-Scl-70-positive patients ($p = 0,009$).

At the baseline average DLco were below normal and were similar in anti-Scl-70-positive and anti-Scl-70-negative patients: $60 \pm 19\%$ и $61,2 \pm 14,3\%$ respectively ($p 0,05$). Within 1 year DLco significantly decreased in all patients, however anti-Scl-70-positive patients demonstrated more pronounced DLco decrease to $53,4 \pm 17,2\%$ ($p 0,0001$), while in anti-Scl-70-negative patients it was less pronounced to $57,2 \pm 12,8\%$ ($p < 0,05$), respectively. Within 5 years significant DLco decrease was found only in anti-Scl-70-positive patients: $53,9 \pm 16,4\%$ ($p < 0,001$).

The time courses of DLco in anti-Scl-70-positive and anti-Scl-70-negative patients by group are shown in Table 1.

Significant DLco decrease over the 5-year period was observed in Group 2 and Group 3, and the lowest DLco ($<50\%$) were observed in Group 3.

Table 1: The time course of DLco in anti-Scl-70-positive and anti-Scl-70-negative patients by group over the 5 year observation period

Observation period	Group 1		Group 2		Group 3	
	With anti-Scl-70 (n = 4)	Without anti-Scl-70 (n = 11)	With anti-Scl-70 (n = 19)	Without anti-Scl-70 (n = 17)	With anti-Scl-70 (n = 17)	Without anti-Scl-70 (n = 3)
In 1 year (M $\pm$ δ)	78.5 $\pm$ 18	61.5 $\pm$ 18	62 $\pm$ 19	64.1 $\pm$ 11	52 $\pm$ 17	64 $\pm$ 1.8
In 5 years (M $\pm$ δ)	70 $\pm$ 12.5	57 $\pm$ 14	56 $\pm$ 14	61.5 $\pm$ 9	45 $\pm$ 16	52 $\pm$ 7
P	> 0.05	> 0.05	< 0.05	> 0.05	< 0.05	> 0.05

Conclusion: Therefore, the presence of anti-Scl-70 antibodies was associated with significantly lower pulmonary function parameters specifically in pts with progressive ILD.

Disclosure of Interests: None declared

DOI: 10.1136/annrheumdis-2019-eular.7932

AB0219 BIOMARKER EXPRESSION IN MONOCYTE SUBPOPULATIONS IN SSC PATIENTS

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Background: Systemic sclerosis (SSc) is a chronic autoimmune disease characterized by vasculopathy and fibrosis, which can be classified into diffuse cutaneous (dSSc) and limited cutaneous (lSSc) subtypes. Monocytes are key agents in the pathophysiology of systemic autoimmune diseases, including systemic sclerosis (SSc). M1 cells (CD14++CD16-) represent a predominantly pro-inflammatory phenotype and M2 cells (CD14-/+CD16+/++) are more associated to a regulatory/pro-fibrotic phenotype.

Objectives: Our aim was to evaluate circulating blood monocyte subpopulations [classical (M1), intermediate and non-classical (M2)] and analyze the expression of CD163, CD169, CD206 and HLA-DR (function and activation monocytes receptors).

Methods: Fifty consecutive patients fulfilling the 2013 ACR/EULAR classification criteria for SSc were included in a cross-sectional study. Monocyte subpopulations were identified and characterized according to the expression of CD64, CD16, CD14, CD163, CD169, CD206 and HLA-DR. Thirty-eight age- and sex-matched healthy individuals were recruited as a control group.

Results: SSc patients mean age was $57,2 \pm 12,8$ years (HC $55,2 \pm 11,4$) and 94% were female. Limited form of disease was present in 72% of SSc patients and. SSc patients had an increased number of circulating peripheral blood monocytes compared to healthy subjects (table 1). Absolute counts of CD16+ (intermediary and non-classical) monocyte subpopulations were higher in SSc patients compared to HC [$79,9$ ($53,4-103,5$)/mm³ vs. $55,9$ ($26,8-85,8$)/mm³, $p=0,003$]. HLA-DR intensity of expression was higher in all monocyte subsets from lSSc and dSSc patients when compared to control. There was a higher percentage of classical [$1,56$ ($0,84-2,98$) vs. $0,68$ ($0,37-1,88$); $p=0,003$] and intermediate monocytes [$15,9$ ($9,5-29,9$) vs. $6,1$ ($3,7-11,5$); $p<0,001$] with CD206 expression in SSc patients compared to HC, and a higher percentage of CD169 expression in dSSc patients compared to control and lSSc groups ($p<0,01$).

Table 1. Absolute value (/mm³) about monocytes subpopulations in all systemic sclerosis (SSc) patients compared to healthy controls (HC) and in limited (lSSc) and diffuse (dSSc) systemic sclerosis subtypes.

Monocytes subpopulations	SSc patients (n=50)	HC (n=38)	p *	lSSc (n=36)	dSSc (n=14)	p * (lSSc x HC)	p * (lSSc x dSSc)	p * (dSSc x HC)
Classical monocytes	346.2 (260.9-450.8)	209.8 (146.1-287.1)	<0.001	363.2 (262.9-451.8)	326.5 (231.2-461.9)	0.779	<0.001	0.003
Intermediate monocytes	38.2 (24.6-47.1)	25.4 (12.6-41.2)	0.005	38.5 (24.7-47.5)	37.1 (22.1-49.1)	0.795	0.009	0.069
Non-classical monocytes	41.2 (20.8-58.3)	28.3 (11.5-46.1)	0.006	42.5 (20.8-69.0)	37.1 (20.8-47.3)	0.310	0.005	0.201

Data are shown as median (interquartile range- IQR). * Mann-Whitney U test

Conclusion: In our study, SSc patients show greater monocytes counts than HC with a predominantly regulatory/pro-fibrotic phenotype (M2). dSSc seems to be more associated to CD169 than lSSc.

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Disclosure of Interests: Laiana Schneider: None declared, Natalia Marcondes: None declared, Vanessa Hax: None declared, Isadora Moreira: None declared, carolina yuka: None declared, rafaela romeiro: None declared, Ricardo Xavier Consultant for: Abbvie, Pfizer, Novartis, Janssen, Lilly, Roche, Rafael Chakr: None declared

DOI: 10.1136/annrheumdis-2019-eular.7890

AB0220

ALPHA- SMOOTH MUSCLE ACTIN EXPRESSION ON ENDOTHELIAL PROGENITORS CELLS OF SYSTEMIC SCLEROSIS PATIENTS: POSSIBLE ROLE IN THE ENDOTHELIAL-TO-MESENCHYMAL TRANSITION PROCESS

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Background: Endothelial-to-mesenchymal transition (EndoMT), a newly recognized type of cellular transdifferentiation, seems to be involved in Systemic Sclerosis (SSc) pathogenesis. In this process endothelial cells lose their specific markers, and acquire a mesenchymal phenotype, thus expressing cell products such as alpha smooth muscle actin (α -SMA) (1,2). Circulating endothelial progenitors cells (EPCs) derive from bone marrow stem cells and contribute to *de novo* vessels formation. Several studies, although with conflicting results, have shown that EPCs in the peripheral blood of patients with SSc are impaired in their number and function (3).

Objectives: to assess the expression of α -SMA, possibly associated with a pro-mesenchymal switch (EndoMT) of the circulating Early (CD34+KDR+CD 133+) and Late (CD34+KDR+) EPCs in the peripheral blood of SSc patients and of patients with Very Early Diagnosis of SSc (VEDOSS) compared with healthy controls (HC) using flow cytometry.

Methods: we enrolled 11 patients (7 SSc and 4 VEDOSS), classified according to the classification criteria for SSc (4) and for VEDOSS not fulfilling SSc criteria (5), and 10 HC. Phenotypic characterization was performed as previously described by Vasa et al. using a FACS Calibur (BD Immunocytometry Systems). EPCs number was expressed as a percentage of cells within the lymphocyte gate.

Results: we found a significantly higher percentage of α -SMA positive Early EPCs (CD34+KDR+CD 133+ α -SMA+) in all patients respect to HC (0,06% $\pm$ 0,03 vs 0,03% $\pm$ 0,01; $p=0,0149$) particularly in VEDOSS patients (0,07% $\pm$ 0,01 vs 0,03 $\pm$ 0,01 $p=0,008$). Moreover, in VEDOSS patients, also the percentage of Early EPCs (CD34+KDR+CD 133+), Late EPCs (CD34+KDR+) and α -SMA positive Late EPCs (CD34+KDR+ α -SMA+) were significantly higher than in HC (0,05% $\pm$ 0,01 vs 0,03% $\pm$ 0,01 $p=0,05$; 0,07% $\pm$ 0,01 vs 0,04% $\pm$ 0,02 $p=0,04$; 0,06% $\pm$ 0,01 vs 0,04% $\pm$ 0,02 $p=0,05$). Besides Early EPCs and α -SMA positive Early EPCs percentages seem to be significantly reduced in patients taking iloprost ($p=0,05$ and $p=0,01$ respectively), calcium channel blockers (CCB) ($p=0,05$ and $p=0,03$) and DMARDs ($p=0,017$ and $p=0,013$).

Conclusion: EndoMT seems to be involved in the pathogenesis of SSc and circulating EPCs seem to be impaired in number and function in SSc patients. In our study we found higher levels of EPCs, in particular α -SMA positive Early EPCs in both groups of patients (SSc and VEDOSS) respect to HC. Thus we can hypothesize a predominant pro-mesenchymal phenotype of this kind of EPCs. This could be considered the expression of the involvement of EPCs in EndoMT process and could better explain the controversial role of EPCs in SSc pathogenesis. Very interesting is the finding of a lower percentage of Early EPCs, and in particular of α -SMA positive Early EPCs, in those patients taking iloprost, CCB and DMARDs, suggesting a potential effect of these drugs on this subgroup of EPCs.

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Disclosure of Interests: Katia Stefanantoni Consultant for: Only 1 scientific advice for Italfarmaco in 2016, cristiana barbati: None declared, Carlotta Angelelli: None declared, Greta Pellegrino: None declared, cristiano alessandri: None declared, Guido Valesini: None declared, Valeria Riccieri: None declared

DOI: 10.1136/annrheumdis-2019-eular.6342

AB0221

SEQUENCE OF CLINICAL SYMPTOMS ONSET AND ITS CORRELATION TO THE AUTOANTIBODIES PRESENCE IN PATIENTS WITH IDIOPATHIC INFLAMMATORY MYOPATHIES

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Background: Idiopathic inflammatory myopathies (IIM) is a heterogeneous group of autoimmune diseases with a broad spectrum of clinical presentations including musculoskeletal, respiratory, neurological, dermatological, gastrointestinal and cardiovascular symptoms. It is known that presence of autoantibodies is a predictor of disease course. However, there is very limited data summarizing long-term observations assessing, when the particular symptoms are expected to occur.

Objectives: To assess the development of the symptoms in IIM patients during the disease course with respect to their serology patterns.

Methods: Medical history of IIM patients treated in our clinic between 2008 and 2019 were reviewed. We excluded patients with: insufficient data for the first period of disease course, history of IIM <6 months, IIM patients who do not fulfill Peter & Bohan 1975 diagnostic criteria (less than 4 points). Finally 103 IIM patients medical history were included for this analysis. EUROLINE test containing 16 myositis specific or associated antibodies (MSAs, MAAs) was used to determine serology for all of those patients. Onsets of symptoms were noted only in case when symptoms were confirmed objectively. Mean time since onset (MTSO) in months was counted for each symptom.

Results: Numbers below are presenting MTSO in months for each symptom.

Anti-Jo1 antibodies positive patients (n:30) have presented: rash (0), mechanic hands (1.2), myocarditis (5), muscle weakness (10.3), Raynaud's phenomenon (RP) (10.4), arthritis (20.7), interstitial lung disease (ILD) (23.5), fever (29.1), Gottron's sign (31.3), muscle pain (46.2), dysphagia (53.4).

Anti-Mi-2 antibodies positive patients (n:12) have presented: RP (0), mechanic hands (0), rash (0.2), Gottron's sign (0.3), fever (2.5), muscle weakness (4.5), dysphagia (4.5), muscle pain (9), ILD (15.5).

Anti-PM-Scl antibodies positive patients (n:7) have presented: rash (1), fever (1), Gottron's sign (1.25), mechanic hands (4), ILD (4), RP (5), arthritis (10.2), muscle weakness (46.3), muscle pain (108.5).

Anti-SRP antibodies positive patients (n:5) have presented: muscle weakness, fever, arthritis and ILD symptoms simultaneously (0), then dysphagia (4) and muscle pain (10).

Anti-NXP2 antibodies positive patients (n:4) have presented: muscle weakness and pain simultaneously with dysphagia (0), ILD developed 4 months later.

Anti-MDA5 antibodies positive patients (n:2) have presented: arthritis (0), fever (0.5), muscle pain (2), rash (3).

Anti-Ro52 antibodies positive patients (n:12) have presented: RP (3), Gottron's sign (3.3), rash (3.5), arthritis (5), muscle weakness (6.2), muscle pain (11), myocarditis (11), ILD (12), mechanic hands (13), dysphagia (18.3).

MSAs and MAAs negative patients (n:21) have presented: fever (0), RP (0), dysphagia (0.3), muscle pain (0.4), muscle weakness (1.9), rash (20), arthritis (66).

Conclusion: Our study shows differences in disease course in IIM patients with each serology pattern adding new data on chronology of appearances of each symptom, which is important in predicting the future course of the disease and planning long-term management.

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Disclosure of Interests: Piotr Szczęsny: None declared, Anna Felis-Giemza: None declared, Katarzyna Swierkocka: None declared, Marzena Olesińska Consultant for: F. Hoffmann-La Roche

DOI: 10.1136/annrheumdis-2019-eular.7758